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Double Nail of the Little Toe

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Key Words
Little toenail · Double nail · Accessory nail · Nail unit resection · Histology

Abstract
A rudimentary accessory or double nail of the little toe is not rare, although only described three times before. Most cases are accidentally detected and only few patients seek help because they have discomfort or pain. Some have a positive family history, but most patients cannot give any information concerning heredity. Clinically, the nail of the little toe is abnormally wide and is split or shows a longitudinal depression corresponding to a slight protuberance of the cuticle. Histopathology shows a complete though short nail. The treatment of choice is segmental excision of the entire accessory nail unit with mobilization of the lateral skin and primary suture.

Introduction

Modern civilization has resulted in poor foot health with many painful conditions and acquired malformations. These are often taken for granted by the affected individuals until they cause discomfort and pain. Often, one condition leads to another and they mutually aggravate each other. Flat and splayfoot are extremely common leading to callus formation in the middle of the forefoot sole, hallux valgus and external rotation of the little toe. Some patients develop a callus or even a corn on the lateral aspect of the little toe that may become painful, particularly when wearing pointed shoes.

In the last 20 years, many patients who had a callus on the lateral aspect of the distal phalanx of the little toe and a peculiar wide little toenail with a split between the lateral and median third were incidentally seen.

Materials and Methods

The author's photographic archive from April 2000 until October 2013 was searched for the diagnosis of 'double nail' or 'double little toenail'. The charts of the patients seen after 2005 were retrieved and evaluated for gender, age, nationality, race, familiarity and treatment. Surgery, either excision or phenolization, was recommended for symptomatic cases. All surgical specimens were examined histopathologically with hematoxylin and eosin and PAS stains. Immunohistochemistry was performed in three cases.

Results

Patients with double little toenails were seen in all countries visited, irrespective of skin complexion and race (table 1). Most cases were detected during an examination for another reason, some patients showed their little toes during foot examination, and only very few consulted because of this condition.

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Table 1. Patients with double little toenails according to year of diagnosis, gender, age at diagnosis, nationality and affected side

Patient	Year	Gender	Age, years	Nationality	Side
TG	2000	F	57	Norwegian	both
NEM	2001	F	47	Norwegian	both
DN	2001	F	42	Norwegian	both
DKZ	2002	F	53	Norwegian	both
HDN	2002	F	49	Norwegian	both
HM	2003	F	37	Norwegian	both
JGM2003	2003	F	42	Norwegian	both
WK	2003	M	43	Norwegian	both
GAB	2003	F	45	Norwegian	both
GDN	2004	M	35	Ghanaian	both
GAM	2005	F	65	German	both
CH	2006	M	60	German	both
DKN	2006	F	38	German	both
DPN	2006	F	42	German	both
HL	2007	M	3 1/2	German	both
FP	2007	M	31	Polish	both
HDB	2008	M	42	Swiss	both
FA	2008	F	41	Italian	both
SB	2008	F	28	German	both
BN	2009	M	62	Swiss	both
DAB	2009	M	38	Swiss	both
SE	2009	F	37	Swiss	both
GP	2009	M	16	Swiss	both
GM	2009	F	60	Swiss	both
RS	2009	F	58	German	left
PG	2009	F	65	German	both
MT	2009	F	59	German	both
WM	2009	M	49	German	both
GD	2009	F	32	Mexican	both
DR	2009	F	28	Swiss	both
FR	2009	F	53	German	both
JWB	2009	F	42	Swiss	both
AW	2009	F	52	Swiss	both (r > l)
GP	2010	F	35	Swiss	both
TN	2010	M	5 1/2	Indian	both
BAMMM	2010	F	36	Portuguese	both
BBM	2010	F	40	Swiss	both
BV	2011	F	59	Swiss	left
GP	2011	M	23	Swiss	both
MB	2011	M	43	Belgian	both
CMAC	2011	F	68	Portuguese	both
BKK	2012	F	47	Thai	both
BKT	2012	F	61	Thai	both
MD	2012	F	13	Swiss	both
SA	2012	F	60	Swiss	both
SD	2012	M	13	Swiss	both
SA	2012	F	22	Swiss	both
SA	2012	F	48	Swiss	both
BT	2013	F	9	Swiss	right
JH	2013	F	45	German	both
SN	2013	M	68	Turkish	both
CL	2013	F	21	Swiss	both
CW	2013	M	48	Han Chinese	both
HD	2013	M	52	German	both
NS	2013	M	62	Tamil	left
BB	2013	F	53	Swiss	left
ZJ	2013	F	36	Swiss	both
DMWC	2013	M	51	Belgian	both

The total number of patients was 58 (20 males and 38 females). The youngest case was 3 1/2 years old and the oldest 62 years. The affected sides of the little toenails were: 1 right, 4 left, and 54 on both sides.

The age of the patients ranged from 5 to 68 years; those individuals that were aware of their condition stated that their toes had always been like this or that they could not remember when it had started. Familial occurrence was only seen when the patient was accompanied by a family member, except for 2 patients with symptomatic double nails who reported that their father and grandfather, or mother and aunt, respectively, also had the same condition.

Clinically, a wide little nail was by far the commonest presentation (fig. 1, 2). Close inspection and dermatoscopy revealed that the nail consisted of two parts with the medial being bigger than the lateral one. They were divided by a longitudinal depression or a real split and the cuticle protruded slightly distally in this particular location. Two really distinct nails were rare (fig. 3, 4). The expression was usually symmetrical on both sides, although roughly one fourth showed a more marked picture on one side.

Discomfort was noted by approximately one third and pain by one fifth of the, mostly elderly, patients. Most of them had a splayfoot with external rotation of the little toe, with an almost vertically growing nail exerting pain (fig. 5). They also often had a circumscribed callus lateral to the nail (fig. 1), which was usually misdiagnosed as a hard corn. Palpation and probing showed that it was the nail, not the callus, that was painful. A radiograph was taken in 10 patients; however, the quality of the X-ray films of the little toe did usually not allow specific details to be detected. Three patients exhibited a lazy Y at the tip of the distal phalanx and 1 a thorn-like bony excrescence. In 2 patients, a Y-shaped tip of the terminal phalangeal bone was identified during surgery.

Fig. 1. Little toes with double nail in a 36-year-old Portuguese woman. Note the callus at the lateral side of the middle interphalangeal joint.

Fig. 2. Both little toes with double nails in a 52-year-old Swiss woman who consulted us for retronychia.

Fig. 3. Asymmetric expression of the double little toenails: two individual right toenails and the typical left double nail adjacent to the main nail.

Fig. 4. A 39-year-old Swiss woman with bilateral double fifth toenails. The left little toe before segmental excision and 4 weeks after surgery is shown.

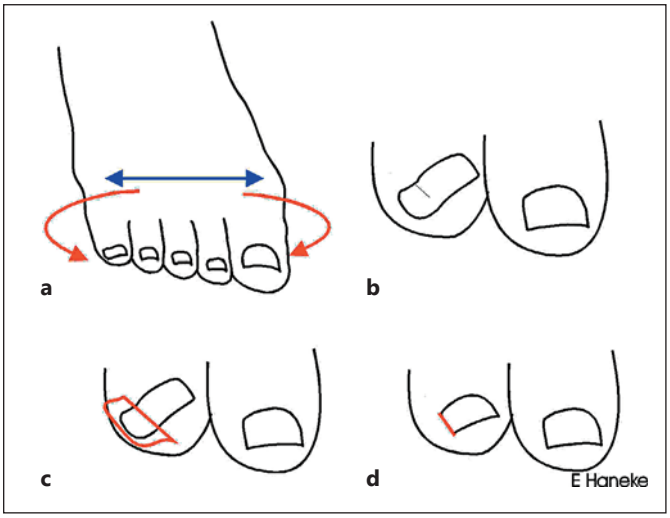

Fig. 5. Schematic illustration of the development of a symptomatic double toe and its treatment. **a** Patients with a splayfoot develop both a hallux valgus with internal as well as external rotation of the little toe. **b** Outward rotation makes the toenail being almost vertically positioned thus exerting symptoms similar to a corn upon pressure. **c** The excision line is drawn for the complete segmental removal of the accessory nail. Like in the big toenail, the lateral matrix horn reaches far proximal-laterally. **d** After the extirpation of the accessory nail, a little toenail of normal width is created.

Most patients did not request treatment. However, for those who accepted treatment, either phenolization of the accessory matrix or complete resection of the double nail was performed. Healing was uneventful in all cases, taking about 10 days for the surgical excision and 3 weeks for the chemocautery. Normal physical activity was taken up 2–3 days after phenolization (fig. 6, 7).

Differential diagnosis comprises traumatic double nail, ectopic nail and nail spiculum after incomplete extirpation of the lateral matrix horn. Histologic examination of the surgical specimens showed virtually all components of a normal nail in the most pronounced cases and only an invagination of a thick hyperkeratotic epidermis with a broad stratum granulosum and a narrow cone of typical nail plate in the center in small double nails (fig. 8). There were all transitions in between these two extremes. The fully developed double nail had a normal or moderately thickened matrix epithelium and a normal though short nail bed with a disproportionately long isthmus. The hyponychium was clearly discernable. The proximal nail fold was well developed, with a relatively thick cuticle and a pronounced true and false eponychi-

Fig. 6. Double nail of the left fifth toe before and 3 weeks after surgical resection of the accessory part of the nail.

Fig. 7. Double nail of the left little toe before and 8 days after phenolization of the accessory nail.

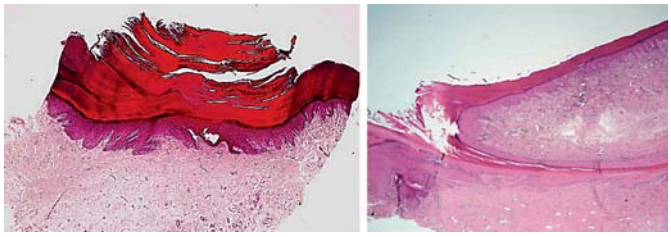

Fig. 8. Histopathology of a poorly developed double nail (left) shows a shallow depression of the epidermis with eosinophilic hyperkeratosis and a pale area in the center representing nail substance, whereas a well-developed double nail (right) shows all characteristics of a normal little toenail with proximal nail fold and thick cuticle, matrix and very short but hyperplastic nail bed.

um. Fungal elements were seen in PAS-stained sections in 1 patient. Immunohistochemistry showed a CD10-positive onychodermis and a weaker CD34 positivity in the matrix and proximal nail bed dermis. Glomus bodies and neural structures were seen as in normal nails.

Discussion

A hereditary dysplasia of the fifth toenail identical to our observations was first published in 1969 [1]. Surprisingly, descriptions of a double little toenail are rare [2, 3], despite its relatively common occurrence. It was thought to be a feature of Han Chinese [3], but among our patients, there was just one Han Chinese colleague who showed his feet during an observational stage. However, double little toenails were observed in dermatological practices in Germany, Norway, the Netherlands, Switzerland, Belgium, Portugal, and Thailand as well as in immigrants from a variety of other countries including Africa (Benin). Thus, this is certainly not a special racial or ethnic feature, although it has to be stated that our results do not allow conclusions to be drawn as to the frequency of double little toenails in different countries, continents and races.

Although rarely stated, the condition appears to be autosomal dominant and occurring in families, both in males and in females. In our study, women predominated, but this is probably due to the fact that they have more often acquired foot problems and wear narrow pointed fashionable shoes. Two mothers came with their healthy children, one daughter and one son, who both had hitherto unnoticed asymptomatic double little toenails like their mothers. However, the expression was variable; whether this is a genetically variable expression or just due to the age is not clear. As most patients did not even mention their double little toenails, it can be assumed that they are not embarrassed by them. In our cases, they were between 2 and 7 mm wide. Symptoms did not necessarily depend on the width of the nail but rather on the severity of accompanying foot anomalies.

Treatment, if requested, is easy. We either performed a selective matrix phenolization using 88% liquefied phenol for a total of 3 times for 1 min under complete anemia with a tourniquet or by segmental excision of the accessory nail including the entire double nail with the matrix, proximal fold and hyponychium. The defect was closed primarily with triple sutures after mobilization of the lateral aspect of the distal phalanx. Triple sutures allow 5–0 suture material to be used as much of the tension is exerted and distributed while tying a single knot.

Pathomechanisms and genetics are interesting aspects to be considered. Fully developed supernumerary digits develop nails. It is known that a bone core has to be present at the distal phalanx for the development of a nail anlage; atelephalangia is associated with lack of nail formation or nail hypoplasia [4, 5], although total complete anonychia also occurs without bone changes [6]. Whether or not an additional bone core, which may be found in the bifid tip of the distal phalanx of the little toe, is sufficient to induce an accessory nail remains a matter of speculation at this moment. On the other hand, nail epithelium

also has a special role in bone formation and digit regeneration. Nail cells were indeed found to influence underlying mesenchymal cells to regenerate digit bone [7].

Another interesting question is how this condition develops. Whether the double little toenail represents the most initial form of a hexadactyly has not been investigated. If it is a rudimentary hexadactyly, it may induce an extra nail anlage. One patient stated that a radiograph taken for a hallux valgus operation had shown a spine-like extension at the lateral aspect of the terminal phalanx of the little toe. Hexadactyly is sometimes hereditary [8]. Another interesting point is that in congenital onychodysplasia of the index finger (Iso-Kikuchi syndrome), also known as COIF, a Y-shaped distal phalanx of the index finger is associated with a defective index nail, such as hemimicronychia and several other types of onychodysplasia [9, 10].

In conclusion, a double or accessory little toenail is a rather common though underdiagnosed and rarely reported condition. It is autosomal dominant with variable expression. Symptoms such as pain and discomfort are infrequent. Histological examination of these double little toenails shows short but otherwise normal nails or less well developed nail structures.

Statement of Ethics

The study was performed in accordance with the Helsinki Declaration and Good Clinical Practice.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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